

# Sponsors

## 3<sup>rd</sup> CNE International Cystinosis Conference Family Conference July 8-10, 2022

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# 3<sup>rd</sup> CNE International Cystinosis Conference

Thursday July 7 <sup>th</sup> , 2022			
Paramedical Meeting			
(Closed session for paramedical professionals and CNE representatives)			
13:00	13:10	Introduction	Fons Sondag
13:10	13:20	Transition protocol from paedetric to adult care	Femke Glaap, Radboud UMC, Nijmegen
13:20	13:35	Discussion	All
13:35	13:45	Therapy compliance	Elke Buit, Radboud UMC, Nijmegen
13:45	14:00	Discussion	All
14:00	14:10	Transition from tube to oral feeding	Ariadne van Hulle, UZ Leuven
14:00	14:15	Discussion	All
14:15	14:35	How to fit the intake of cysteamine with the food & drinks schedule: Procysbi	Round table discussion
14:35	14:50	Tea break	Room 00.03

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Thursday July 7<sup>th</sup>, 2022

## Paramedical Meeting

(Closed session for paramedical professionals and CNE representatives)

14:50	15:00	Early enteral support in children with cystinosis: impact on growth.	Carmen Campana
15:00	15:15	Discussion	All
15:15	15:25	Nutritional Care in Cystinosis	Judith Erler
15:25	15:40	Discussion	All
15:40	16:40	Wrap-up: record the results of the meeting	All
16:40	16:55	Participants appreciation of the meeting	All
16:55	17:00	Closure	Fons Sondag

# Discussion

## Transition from paedetric to adult care

- Nijmegen
  - Presentation Femke Glaap-Roeven
  - How do you do transition in your hospital?
  - Nijmegen questionnaire: 12-16 and 16-18 ready steady go
- Rosenheim:
  - Doctors do the transition
  - Local hospitals
  - SAVE program: dietic, speech, etc. recommendations summary send to local hospital.
  - No protocol like in Nijmegen
  - Rosenheim has psychologist helping with transition
- Hanover
  - Rehab week in Austria for transplant patients
  - 16-18, 16 years is in fact too late to start
- Leuven
  - Patients stay in same building
  - Ready steady go
  - Stays in paediatric till transplanted
  - If transplanted move to adult nephrology
  - Patients move from Elena Levtchenko to David Cassiman
- Are parents also involved in transition:
  - Nijmegen: Yes, trained to go from caregiver to coach also questionnaire

# Discussion

## Treatment Compliance

- Nijmegen
  - Presentation Elke Buit
  - Knowing your patients, have connection, what do they do, have a relation?
  - Health believe model
  - Chronic disease: never stops
  - Vitamin B12 to avoid smell
- Rome
  - Difficult balance between medication and food, salty food, nausea
- Rosenheim
  - Take lower doses or nothing, because of the smell in morning, or when they go out.
  - They use shots to avoid: medicines mixed with grinded vegetables like spinach to avoid odour
  - One pill is no problem for the stomach, but with a large number of pills they will stay in the stomach
  - Do not want to take pills in in front of their peers
- Leuven
  - Smell biggest problem for adolescents
  - You have to understand their struggle, scared to take medicines with friends, psychologist even went to school to stop bullying. Intensive interaction every 2 weeks
- Should you tell how severe the disease is?
  - No for young children
  - Adult patients: stay positive

# Discussion

## Transition from tube to oral feeding

- Leuven
  - Will be presented at a later stage and not in this meeting
  - Not discussed

# Discussion

## How to fit the intake of cysteamine with the food & drinks schedule: Procysbi

- Leuven
  - Presentation Katrien Van der Vaerent & Ariadne Van Hulle
  - Oral: Open Procysbi capsules, put in fruit juice, take it to school in a bottle
  - Micky Button: mix of apple juice & apple sauce
- Rosenheim
  - Pharmacy looks what medicines can be put together with the nephrologist for a specific patient. Put in blister or in syringes.
  - More smell with Procysby. Use lower dose in the morning to avoid and take more pills later.
  - Not clear what the effect on cystine level is. Dieticians do not see that.
- Idea from the meeting:
  - Cystagon in the morning, cystagon in the afternoon, procysbi for the night or
  - Procysbi 3 times a day
- Rome
  - Avoid high fat and protein. Combine with light meal in the morning
- Parents:
  - Use big syringe where they can mix Procysbi with orange juice, push fast in the gastric tube
  - Get Procysbi granulates from the capsules and put them in juice and then drink it
  - Do not eat 1 hour before and after taking the Procysbi: the most difficult in the morning
  - Adults eat and take Procysbi generally at 10 and 22 hours

# Wrap-up

## Early enteral support in children with cystinosis: impact on growth

- Rome
  - Presentation Carmen Campana
  - Follow infant patient every month
- Nijmegen
  - Follow-up: Once a quarter < 5year
  - Follow-up: Once a year >5 year
  - Tube feeding as extra, e.g. during the night
- Hanover
  - Follow-up: Every quarter
- Leuven
  - Tube feeding with formula, less risk on infection
  - Tube feeding is in most cases supplementary
- Rosenheim (speech therapist)
  - Let them smell, see and taste food and eat normal food
  - People with tube feeding end up with problems to be solved by speech therapist



# Wrap-up

## Nutritional Care in Cystinosis

- University of Plymouth Judith
  - Presentation by Judith Erler
  - Very little research on cystinosis and diets.
  - Current dietic treatment by interviewing dieticians
  - QoL and diet cannot be separated.
- Nijmegen
  - Dietary needs for adult patients? Swallowing becomes a problem, slower eating,

# Participants appreciation of the meeting

- Parent: interesting learn of each other
- Parent: privilege, still learning
- Dietician Rome: learn to know other points of view also from other disciplines
- Physiotherapist Rosenheim: only one in the meeting, should have more than one in next conference
- Speech therapist Rosenheim: only one in the meeting, great first meeting, name the specific therapists you want to include in the meeting in the invitation, register profession in registration
- Dietician Hanover: interesting meeting, how will medication intake be in the future, more combined?
- Medical assistant blood analysis Rosenheim: bring all the points of view from different disciplines together.
- Judith: Great afternoon, listen to the others, new ideas, everyone having the same problems good sharing
- Specialised nurse Nijmegen: Valuable that there are parents of patients of patients in the meeting. Invite also adult patients in the next meeting.
- Specialised nurse Nijmegen: Really appreciate interaction, exchange of experience
- Dietician Nijmegen: Surprisingly good that we had many discussions. Do not spent too much time on presentations, the discussion is more important.
- Dietician Nijmegen: learned how other hospital work with the medication. Also learned from the parents. Ask this group for preparing meeting next conference.

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## Closure of the 1<sup>st</sup> Paramedical Meeting

I want to thank  
all of you for participating  
in this 1st Cystinosis Paramedical Meeting

Fons Sondag, Conference Organisation

